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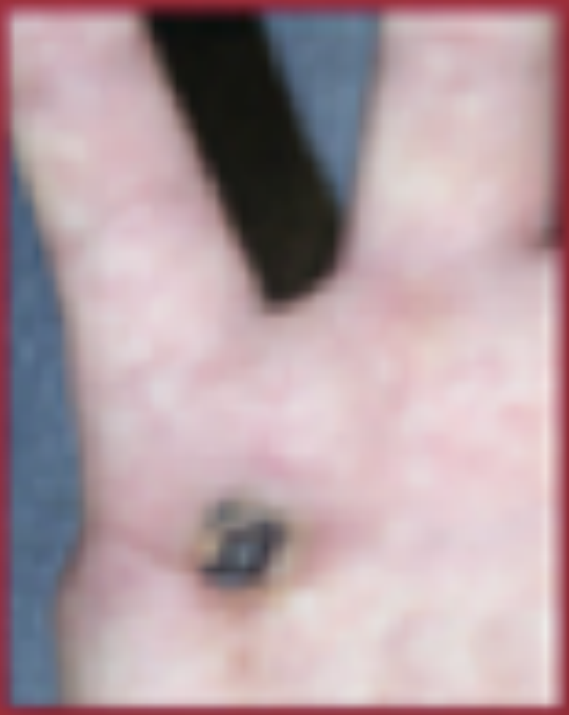

Epithelioid Sarcoma Resembling Benign Fibrous Histiocytoma

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Practice Points

- Epithelioid sarcoma should be considered in the clinical differential diagnosis of nonhealing recurrent lesions of the distal extremities in a young adult.
- Histological presentation of epithelioid sarcoma can mimic a number of benign granulomatous and fibrohistiocytic processes, including benign fibrous histiocytoma.
- Deeper biopsies may be needed to demonstrate the overtly malignant morphology characteristic of epithelioid sarcoma.
- Inactivation of SMARCB1/INI1 is a common molecular aberration identified in epithelioid sarcoma and can be demonstrated immunohistochemically by absence of nuclear staining in tumor cells.

Epithelioid sarcoma (ES) is a rare malignancy notorious for its tendency to histologically mimic granuloma annulare and other palisading granulomatous processes. We report a case of ES on the right hand of a 23-year-old man that histopathologically resembled a benign fibrous histiocytoma. Superficial portions of the tumor were well differentiated, exhibiting spindled and ovoid cells with scant cytoplasm that surrounded sclerotic collagen bundles. More obvious atypia including greater cellularity, nuclear pleomorphism, and mitotic activity were mostly confined to the deep-seated regions of the tumor. In addition to palisading granulomatous processes, ES can mimic benign fibrous histiocytoma, and the superficial portions of ES may appear deceptively benign.

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Epithelioid sarcoma (ES) is a rare malignant soft tissue neoplasm that is most often encountered on the distal extremities of young adults.[1] Epithelioid sarcoma is notorious for its tendency to mimic palisading granulomatous processes such as granuloma annulare. We report a case of ES on the right hand of a 23-year-old man that resembled a benign fibrous histiocytoma (dermatofibroma) on incisional biopsy. The typical histopathologic features of ES were identified after amputation of the hand and evaluation of the deeper regions of the tumor. The tendency for ES to mimic granulomatous processes is a common diagnostic pitfall, but the potential for its close resemblance to benign fibrous histiocytoma is less recognized.

Case Report

A 23-year-old man presented with a nonhealing lesion on the right palm. His medical history was remarkable for a giant cell tumor of the tendon sheath involving the right fifth finger that had been treated via excision at an outside institution 2 years prior. Clinical examination revealed a 0.8×0.6-cm painful, firm, ulcerated dermal nodule with a hemorrhagic crust on the palmar surface of the right hand (Figure 1A). The clinical differential diagnosis included melanoma, traumatized verruca vulgaris, thrombosed pyogenic granuloma, and foreign body. A shave biopsy demonstrated verrucous epidermal hyperplasia, but the specimen did not include the dermis. Cultures of the lesion were positive for *Staphylococcus aureus*, and antibiotic therapy was initiated. In light of the clinical findings and the patient's history of a giant cell tumor, imaging studies were performed. Magnetic resonance angiography showed abnormal masslike infiltrative enhancement throughout

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The authors report no conflict of interest.

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